Cutaneous Manifestations of Connective Tissue Disease - II

Connective tissue diseases

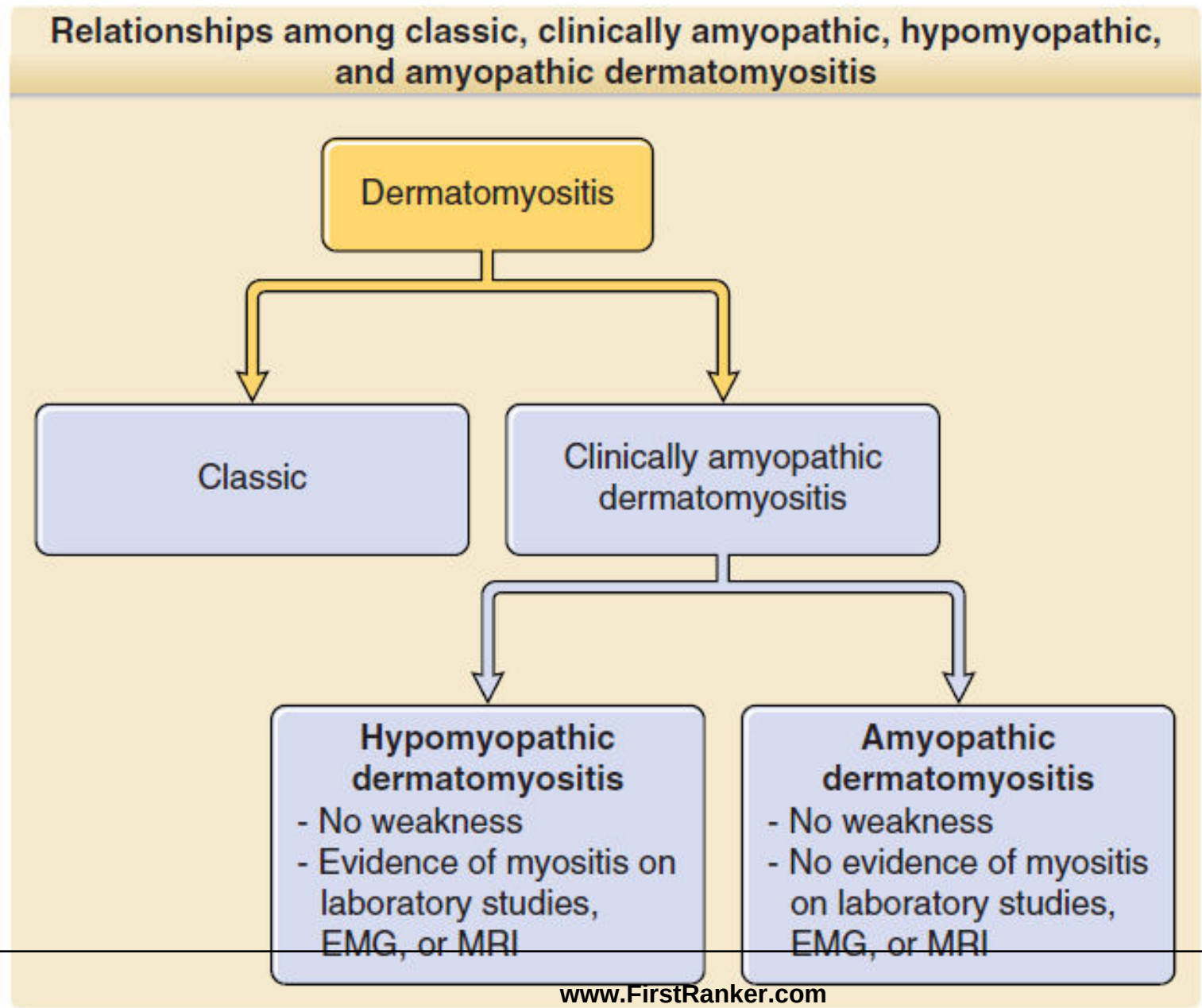
- SLE
- Dermatomyositis
- Systemic sclerosis (scleroderma)
- Rheumatoid arthritis
- Mixed CT disease
- others

- Spectrum ranging from benign cutaneous variants to severe, often fatal, multisystem diseases
- Inflammation of the connective tissue - resulting in changes in skin, joints, vasculature and other Organs
- Antibodies formed against cell components - Serological markers

Dermatomyositis

- Dermatomyositis (DM) - systemic autoimmune disease characterized by inflammation and damage to the skin and muscle
- Interstitial lung disease (ILD) - 20%
- In adults - DM - heralds the diagnosis of a coexisting internal malignancy in 10% to 20% of cases

- Characteristic autoantibodies
  - Antisynthetase
  - Anti–Mi-2
  - Anti–transcriptional intermediary factor [TIF1]-gamma
  - Anti–melanoma differentiation–associated gene 5 [MDA5]
  - Anti–nuclear matrix protein 2 [NXP2]
  - Anti–small ubiquitin-like modifier activating enzyme [SAE]
- May be useful in identifying clinical subsets

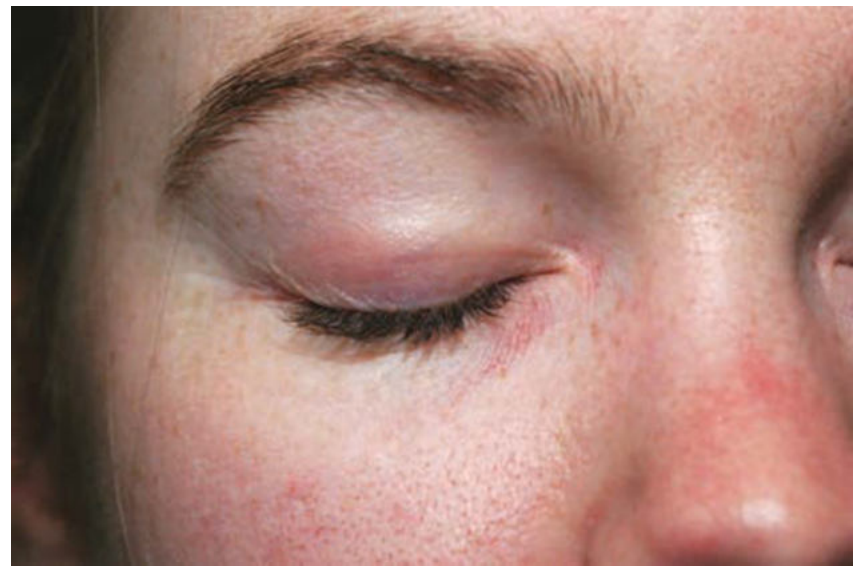


Violaceous patches and plaques

- Characteristic cutaneous feature
- violaceous patches and plaques, varying from a bright pink to a deep violet color

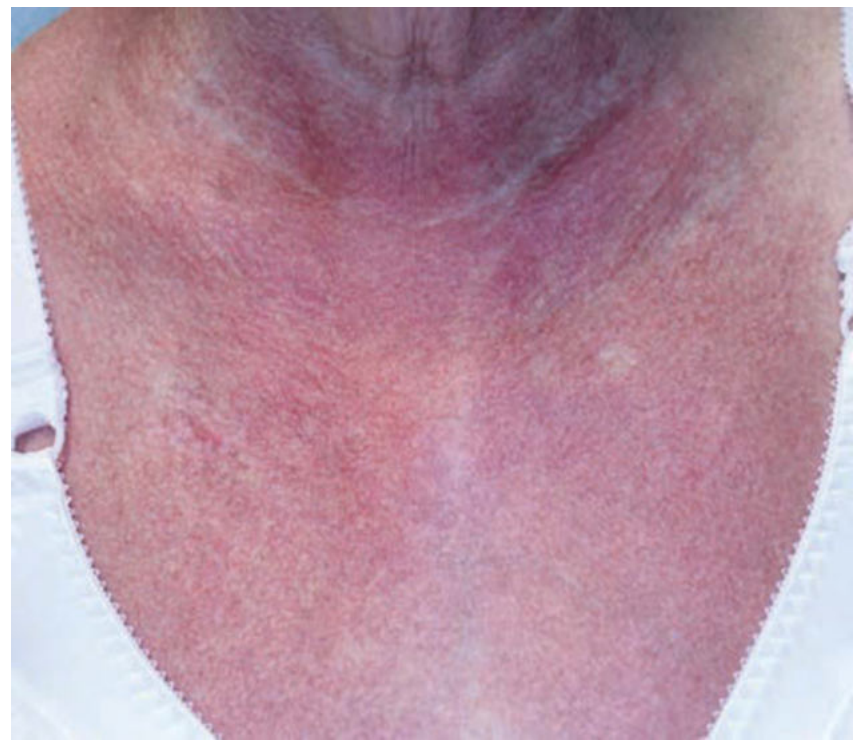
The heliotrope sign

- Exemplies the pink to purple violet hue of the eruption
- color of the flower petals after which the sign named
- The eyelid eruption can be associated with periorbital edema

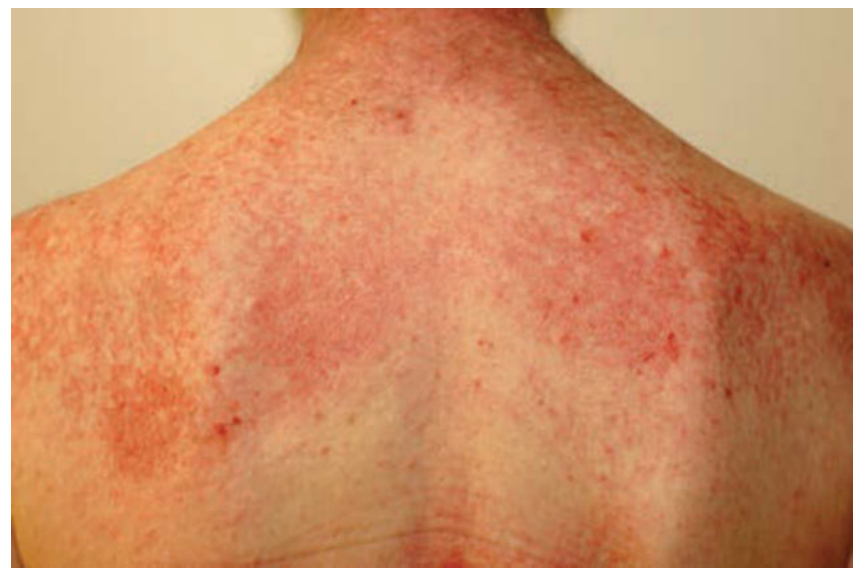


The V-neck sign

- Confluent violaceous erythema on the sun-exposed areas of the lower anterior neck and anterior chest

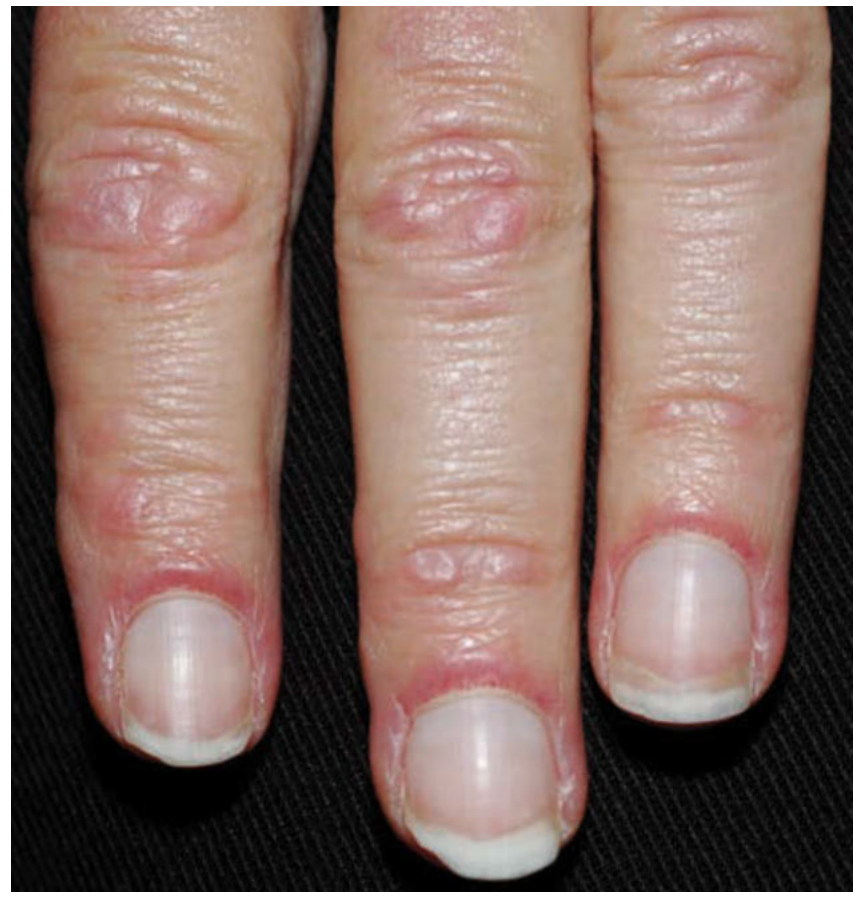


- The skin changes - often distributed to typical regions on the body
- Trunk involvement is often seen on the posterior neck, upper back, and shoulders, known as the shawl sign



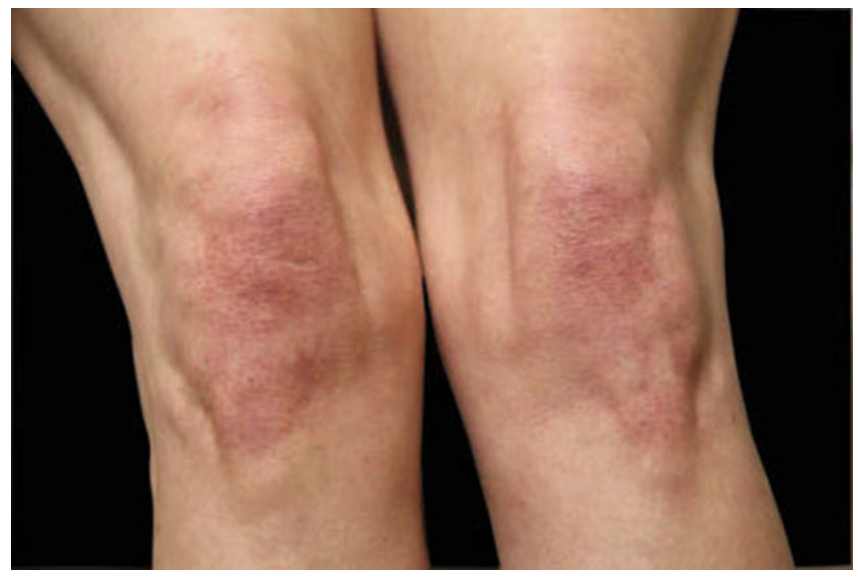
Gottron papules

- The violaceous to pink papules over the IP and MCP joints



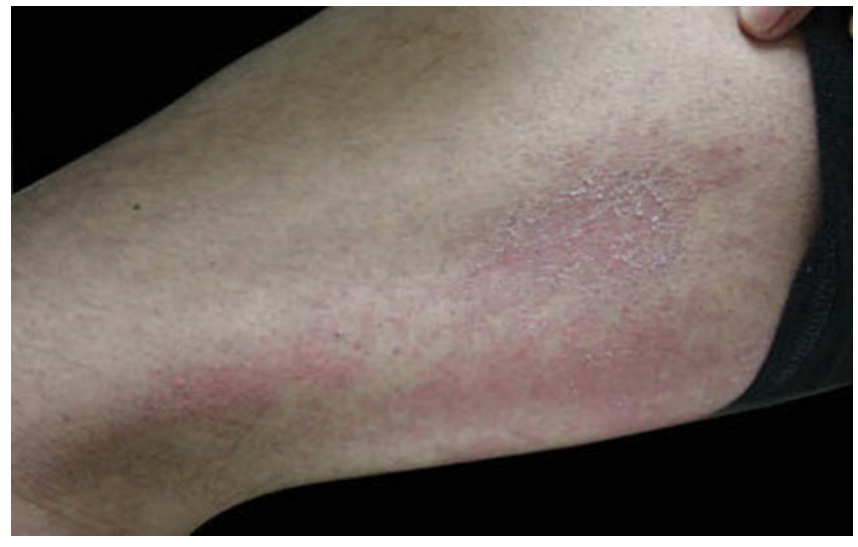
Gottron sign

- Symmetric macular violaceous erythema over the IP joints, olecranon processes, patellae, and medial malleoli
- May be atrophy and poikiloderma in classic areas of Gottron sign



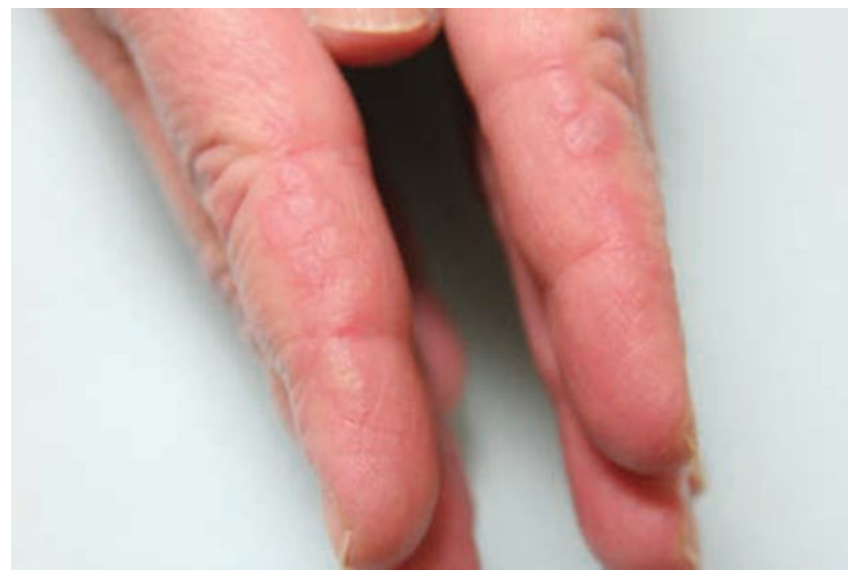
The Holster sign

- The violaceous erythema and poikiloderma on the lateral hips and lateral thighs
- Often patterned as folliculocentric macules or subtle papules



“Mechanic’s” hands

- Hyperkeratosis and fissuring along the medial thumb and lateral second and third digits
- A cutaneous clue to the possible presence of ILD



Systemic sclerosis (Scleroderma)

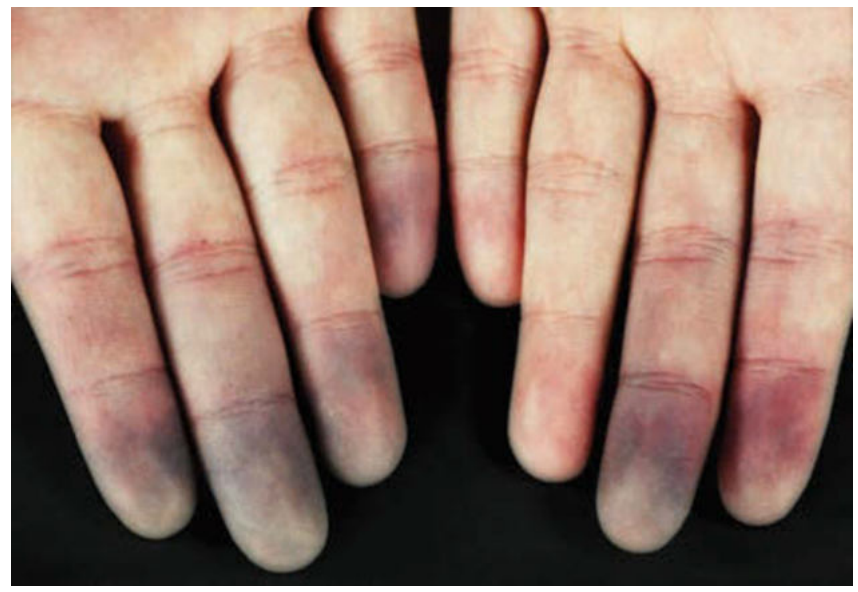
- Multisystemic autoimmune disease - by vasculopathy, inflammation, and fibrosis of the skin and many other organs due to extensive activation of fibroblasts
- Raynaud phenomenon, circulating autoantibodies, and skin sclerosis are almost always present
- Important for the early diagnosis

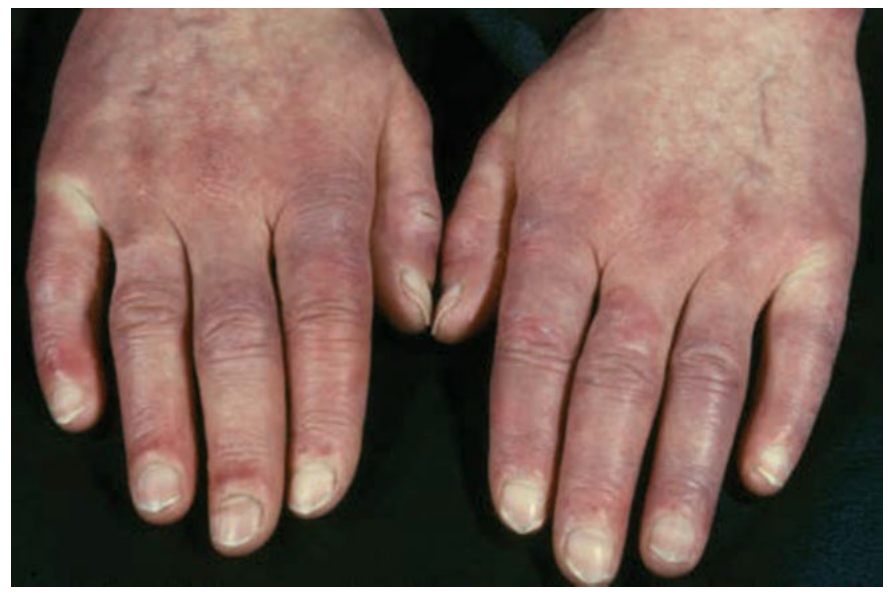
- An overlap syndrome, including mixed connective tissue disease – characterized by additional clinical features of other RHDs
- The highest case-specific mortality of any of the autoimmune rheumatic diseases

- SSc usually starts with a Raynaud phenomenon - precede the disease for many years
- The clinical manifestations - diverse with severe fibrosis of the skin and all additional cutaneous manifestations
- E.g. hardening of the skin, development of contractures, digital ulcerations and calcifications
- Multiple patterns of internal organ involvement

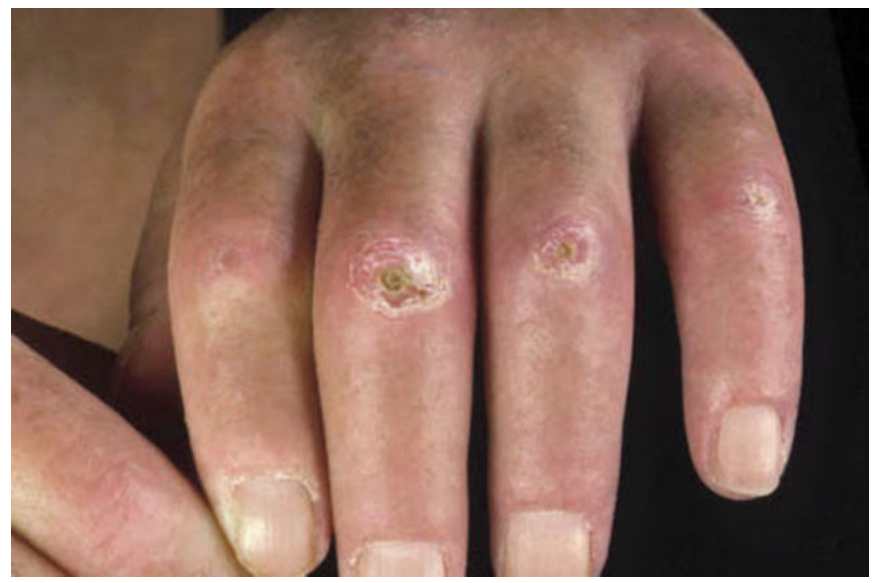
Raynaud's phenomenon

- Earliest symptom to appear in more than 90% of SSc patients
- Characterized by painful pallor/ischemia of single or several digits followed by reactive hyperemia after reheating at the end of a RP attack f/b in some cases cyanosis (triphasic RP)
- Worse in winter/by emotional stress

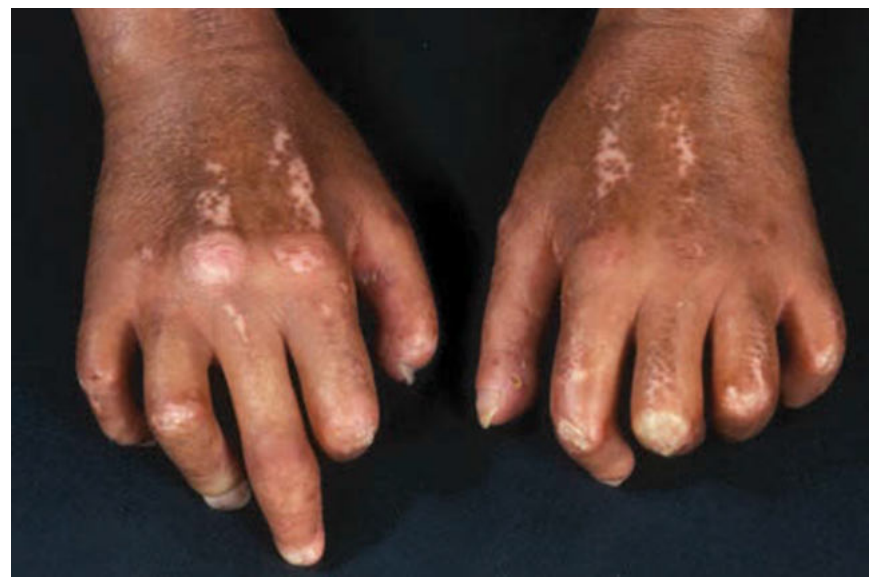


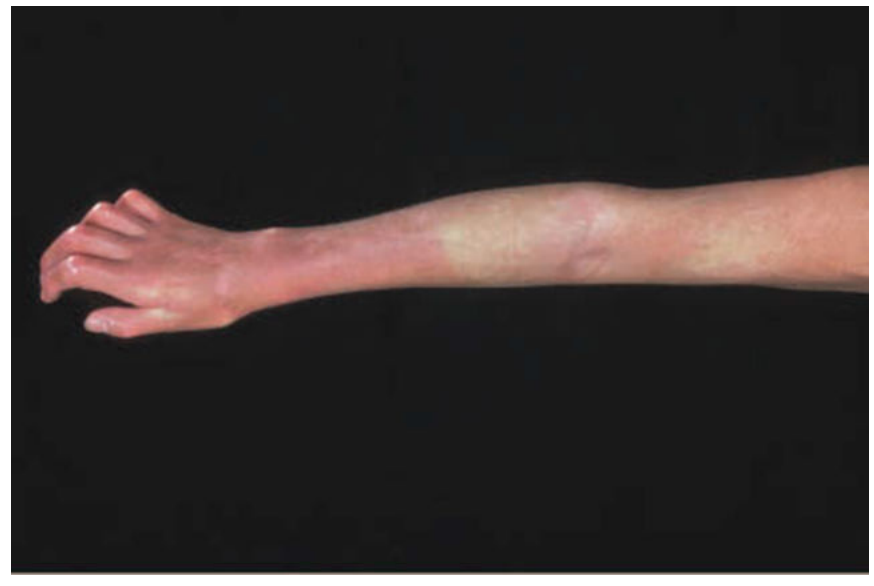


- Eventually leads to:
  - ➢ Finger tip ulcers
  - ➢ Loss of finger pulp
  - ➢ Gangrene

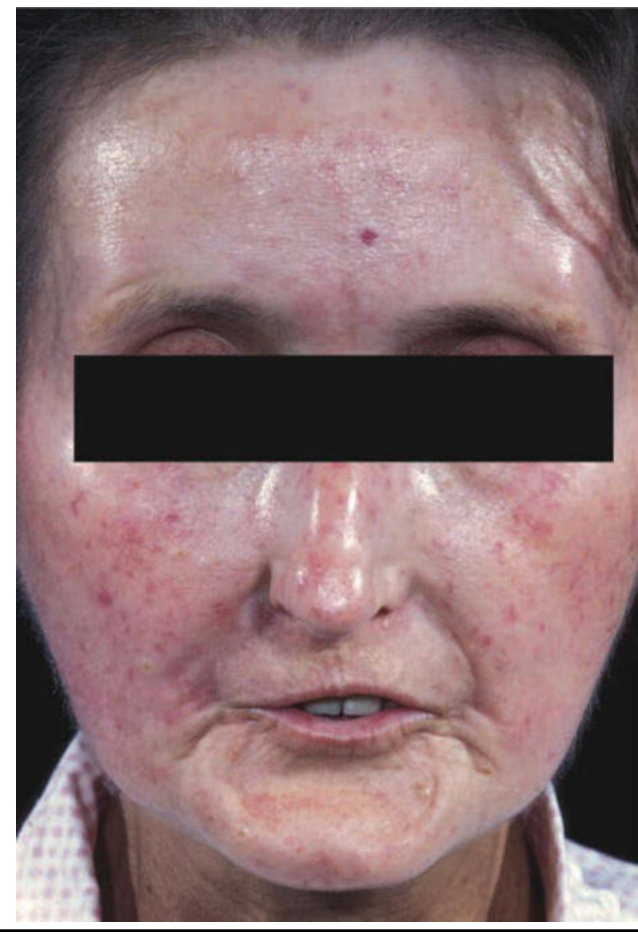


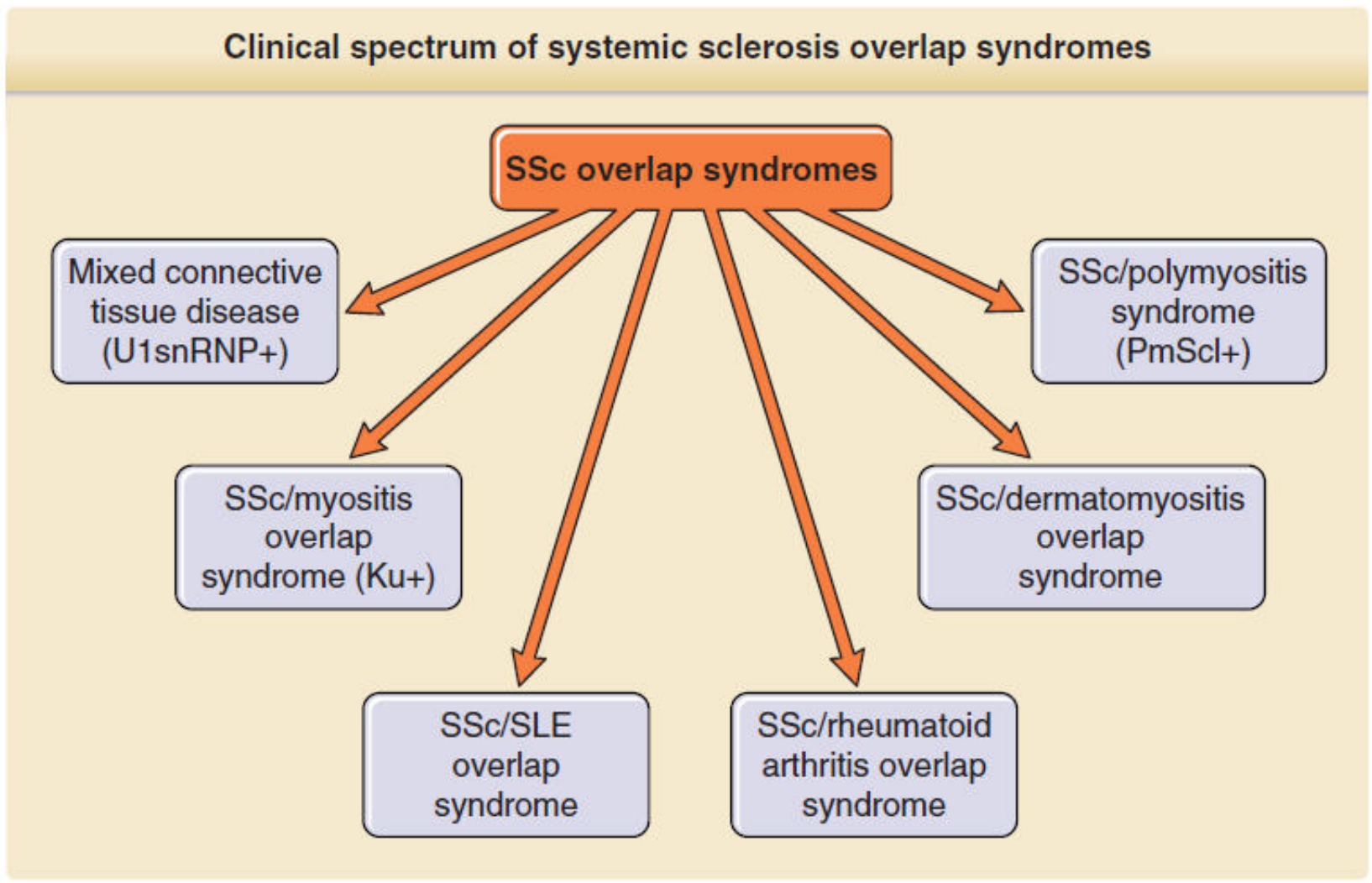
- An increasing induration and skin thickening (sclerodactyly)
- Depending on the localization of skin thickening restricted mobility of joints (dermatogenous contractures)





- Facial appearance - characterized by a radial furrowing around the mouth, no expression, a stiff and mask-like facial appearance, and sclerosis of the frenulum
- Besides cosmetic/aesthetic problems, this causes considerable difficulties regarding eating and oral hygiene





Clinical Features of Mixed Connective Tissue Disease[17,18]

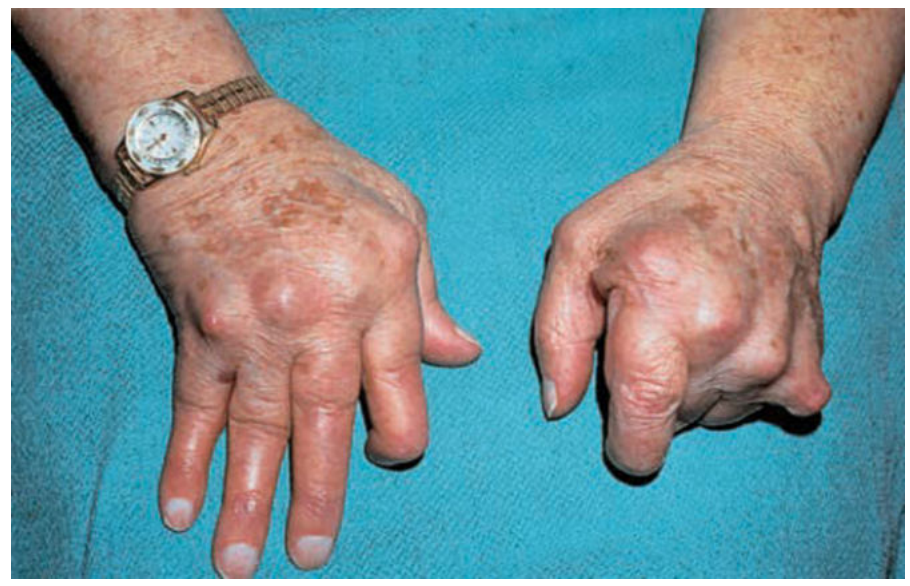
- Raynaud phenomenon
- Puffy fingers/hands
- Sclerodactyly
- Oesophageal involvement
- Pulmonary hypertension/interstitial lung disease
- Myositis
- Arthritis and arthralgia
- Serositis
- Anemia/lymphopenia
- High titers of U1RNP antibodies

Rheumatoid Arthritis

- Affects roughly 1% of the world population
- Skin findings quite varied, including papules, plaques, and nodules with multiple histologic types, vasculitis/Bywaters lesions, pyoderma gangrenosum/Felty ulcers

Rheumatoid Nodules and Nodulosis

- The usual location is over pressure points such as the olecranon, the extensor surface of the forearms, and the Achilles tendon
- Benign, they can lead to complications, including ulceration, infection, joint effusion (rheumatoid chyliform bursitis), and fistulas (fistulous rheumatism)



- Accelerated nodulosis- Low-dose methotrexate, often used for the treatment of RA, may precipitate erythema in and enlargement of preexisting rheumatoid nodules

Rheumatoid neutrophilic dermatosis

- Rare cutaneous manifestation
- Lesions are usually chronic, erythematous, and urticaria-like plaques and papules; sharply marginated